BRIEF REPORT

Anxiety Disorder in a Case of Arnold-Chiari Malformation

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This report describes a patient who suffered concurrently from panic disorder with agoraphobia and Arnold-Chiari malformation. Surgical correction of the neuroanatomical anomaly altered the patient's symptom pattern, enabling a more clear delineation of her anxiety disorder.

Key Words: anxiety disorder, Arnold Chiari, panic, agoraphobia, corrective surgery

INTRODUCTION

Arnold-Chiari malformation is a congenital anomaly leading to herniation of the cerebellar tonsils below the level of the foramen magnum. In the Type I malformation there is caudal descent of the cerebellar tonsils only, while in Type II the cerebellar vermis and possibly the fourth ventricle and pons are involved.

The anomaly may present in a variety of ways (Eisenstat et al 1986) and at times with vague symptoms (Susman et al 1989). Diagnosis is often difficult and therefore often delayed (Eisenstat et al 1986; Raynor 1986). Common symptoms in order of frequency include pain (headache, chest pain, neck pain, arm and leg pain), weakness in arms or legs, sensory loss, ataxia, vertigo and lower cranial nerve abnormalities. To date only one case has been reported of an anxiety disorder in association with this anomaly (Iwabuchi et al 1985).

Case report

Mrs. WK is a 34-year-old widow with two children. She had previously worked as a nursery school teacher. Since the age of ten, the patient had experienced headaches, ataxia and syncope with palpitations. The headaches were usually precipitated by a Valsalva maneuver and were often accompanied by dizziness. Mrs. WK received various treatments for these symptoms over 24 years. She had her right first rib removed, numerous treatments for migraines and a tooth extraction because of possible nerve involvement. She also received holter monitoring for heart palpitations. At the age of 30, Mrs. WK began to experience infrequent episodes of dizziness, unaccompanied by Valsalva maneuvers, and anxiety that she patient described as an "inside tremor." Two years later her husband died, and she began to experience these episodes more intensely. These episodes included symptoms of dizziness, hand tremors, palpitations, chest tightness, hot flashes and a cold, clammy feeling. The symptoms would develop suddenly and unexpectedly and increase in intensity within several minutes. Although these attacks were infrequent, each one was followed by a persistent fear, which lasted for at least one month, of having another attack. The patient avoided shopping and various appointments for fear that these attacks would overwhelm her. She was diagnosed by her family doctor as suffering from panic disorder with agoraphobia. She was treated by a psychologist with weekly group sessions and lorazepam, up to 6 mg/day. The

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patient eventually discontinued the group sessions, and lorazepam was reduced to 0.75 mg/daily because of the patient's concerns about addiction. She benefitted little from these treatments.

One year later she was seen by a neurologist, who noted wide-based gait and ataxia. She exhibited no other motor, sensory or cranial nerve abnormalities. Magnetic resonance imaging (MRI) revealed a Type I Arnold-Chiari malformation. A suboccipital decompression was performed to correct the anomaly.

After surgery Mrs. WK's ataxia resolved, and she no longer experienced syncope with Valsalva maneuvers. However, she continued to experience symptoms consistent with panic disorder with agoraphobia.

She was readmitted to hospital for further investigation. Cushing's disease, pheochromocytoma, thyroid disease and Wilson's disease were ruled out. A repeat MRI of her cervical spine, EEG and EKG were all normal. An ENT consultation ruled out vestibular pathology. The patient was not reassured by this extensive investigation. She felt that her doctors were missing the real cause, since they had missed the diagnosis of Arnold-Chiari malformation for many years.

It was at this point that a psychiatric consultation was requested. Mrs. WK recalled that, as a child, she had been prone to anxiety, even before the onset of her symptoms of an Arnold-Chiari malformation. She dreaded being alone and frequently missed school because of abdominal pain. Because of her somatic complaints, she felt her mother overprotected her. In later years, she became dependent on her husband and extremely anxious during his periodic absences. Her maternal uncle had been hospitalized for treatment of anxiety disorder.

Mrs. WK was diagnosed as having panic disorder. Clonazepam, 0.25 mg four times daily, and individual psychotherapy were started. This was supplemented by a program of desensitization and relaxation. The patient improved significantly. Her agoraphobic symptoms disappeared, and although she continued to experience episodes of dizziness and mild anxiety, their intensity was greatly lessened and she no longer met the DSM-III-R criteria for panic disorder.

DISCUSSION

To our knowledge, there is only one previous report of anxiety disorder in association with Arnold-Chiari malformation. Iwabuchi et al (1985) described a 30-year-old Japanese woman who had a five-year history of insomnia with choked feelings, palpitations, clumsiness of hands and anxiety. She experienced a sudden respiratory arrest related to sleep apnea, and Arnold-Chiari malformation was eventually diagnosed.

In the case described here, a family history of anxiety suggests a genetic predisposition. There is early evidence of a predisposition to an anxiety disorder from her childhood history. Her mother's overprotectiveness may have hindered her separation-individuation process and contributed to the perpetuation of her anxiety-prone nature. Symptoms of Arnold-Chiari malformation may have acted as a trigger for episodes of anxiety. One might speculate that the neuroanatomical anomaly led to a greater prevalence of symptoms which could engender anxiety or to the subjective feeling of anxiety in a patient so predisposed. Alternatively, abnormal pressures on the medulla and pons may directly induce anxiety symptoms. An increase in anxiety symptoms has been found with other neurological disorders. For example, limbic epilepsy seems to predispose a person to anxiety. Adamec (1990) demonstrated that kindling of the rat amygdala to produce seizures also increased their anxiety response.

Patients tend to do poorly after corrective surgery for an Arnold-Chiari malformation if their symptoms have persisted more than 24 months (Dyste et al 1989). A history of chronicity may account in part for the continuation of this patient's symptoms. However, the change in pattern of symptoms seems to have resulted largely from the elimination of symptoms arising from the neuroanatomical anomaly. Once this change occurred, her anxiety disorder could be more clearly delineated, allowing for appropriate treatment and the alleviation of the symptoms. This case illustrates how the presence of a psychiatric syndrome in patients with overlapping neurological disease can delay diagnosis and therefore appropriate treatment for both conditions.

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